Case Presentation and Review: Constrictive Pericarditis

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CONSTRICTIVE PERICARDITIS (CP) is present when a thickened and adherent pericardium restricts diastolic filling of the heart. Tuberculosis was once a common cause of CP, whereas nowadays most cases are idiopathic.[1] Previously known as Pick's Disease, CP often is referred to as pseudocirrhosis because it clinically can mimic chronic liver disease.[2,3] Patients with CP frequently have hepatomegaly, marked pitting lower extremity edema, and ascites with liver function tests indicative of passive congestion. The key differentiation from liver disease is the elevated jugular venous pressure. The possible presence of Kussmaul's sign and the pericardial knock also help distinguish CP. Echo-Doppler cardiography, a diagnostic tool, excludes other diseases and has the superior ability to evaluate diastolic flow and function. Patients suspected of having CP should undergo right heart catheterization in order to document equalization and elevation of filling pressures. With the improvement of imaging techniques, the diagnosis can be supported by CT or MRI scanning with a high degree of sensitivity. The dramatic improvement resulting from pericardial stripping in patients who have not progressed to advanced stages of heart failure points to the importance of recognizing this uncommon yet potentially curable form of heart failure.

Clinical Presentation

A 48-year-old man was referred to a clinic with a three-year history of anasarca and lower extremity edema which were partially responsive to the medications lasix and spironolactone. The patient was told his symptoms were partially a result of his history of alcohol abuse. Although he had suffered chronic obstructive lung disease with an FEV1 of 66% of normal, he had not had tuberculosis and was PPD negative. His medications were lasix 60 mg. BID and spironolactone 50 mg. QD. On physical examination his blood pressure was 115/63; pulse, 87; respirations, 18; and he was not febrile. There was a pulsus paradoxus of 15 mmHg. He had no spider angioma or icterus. His neck veins were distended, and his lungs were clear to auscultation. On cardiac exam he had a regular rhythm, with a soft I/VI systolic murmur at the apex. He had an enlarged liver percussed at 20 cm, a fluid wave with bulging flanks, and marked pitting edema to his knees bilaterally (Table 1).

The patient was referred to cardiology for a right and left heart catheterization and biopsy. Right heart catheterization was consistent with diastolic equalization of pressures (Figure 1), and hemodynamic representation of Kussmaul's sign was seen (Figure 2). Simultaneous right and left heart catheterization showed elevation and equalization of right sided and left sided diastolic filling pressures, with characteristic dip and plateau (Figures 3 and 4). The patient's arterial tracing demonstrated a pulsus paradoxus of 15 mmHg (Figure 5). No evidence of left to right shunt by sampling venous blood in the right heart chambers was noted.

The patient underwent non contrast computed tomography of the chest which confirmed a thickened pericardium of 5 mm (normal <4 mm), and pericardial stripping resulted in marked resolution of his symptoms.

This case demonstrates the clinical similarities of CP and chronic liver disease, yet also points out features that distinguish the two. Most importantly, the jugular venous pressure (JVP) of the CP patient is elevated, but is usually normal in patients with cirrhosis. In fact, astute observation reveals a sudden collapse of the JVP (rapid y-descent) in patients with CP (Figure 2). Furthermore, patients with long-standing cirrhosis usually have small nodular livers, and patients with CP frequently have enlarged, smooth livers. The liver function tests in patients with CP may have a pattern of passive congestion with more prominent elevations of alkaline phosphatase and GGT than transaminases, and cirrhotic patients often have more diminished synthetic function with decreased albumen and elevated protime and, possibly, elevated transaminases (Table 2). Because of CP chronicity, the finding of decreased liver synthetic function with a depressed serum albumen makes the liver function tests poor for distinguishing CP from chronic liver disease.

Although an elevated JVP can help distinguish patients with CP from patients with cirrhosis, the conditions of cardiac tamponade, right ventricular infarction, and restrictive cardiomyopathy all can produce an elevated JVP and appear clinically similar to CP. A rapid y-descent of the JVP supports the diagnosis of CP. In addition, the pericardial knock is only found in CP, but it is heard in just one-third to one-half of cases.[4] The pericardial knock, heard best with the diaphragm of the stethoscope, occurs 0.6-0.12 seconds after S2—not to be confused with an S3, which is of lower frequency—and corresponds to the rapid cessation of ventricular filling.[4] Given the variable degree in which various physical findings are recorded in the literature (Table 3),[2,3,5-7] one is often left with the differential diagnosis of CP, tamponade, and restrictive cardiomyopathy when embarking on the diagnostic workup.

Introduction

Constrictive pericarditis, defined as impedance to diastolic filling caused by a fibrotic pericardium, may be a

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ABBREVIATIONS USED IN TEXT

JVP = jugular venous pressure
MRI = magnetic resonance imaging
CP = constrictive pericarditis
CT = computed tomography

long-term consequence of chronic pericarditis following an acute episode. The visceral and parietal pericardium usually become adherent with obliteration of the pericardial space, but in some cases the constricting process is formed by the visceral pericardium (epicardium) alone.[5,8] As the disease progresses, calcium deposition may occur with further thickening and stiffening of the pericardium. Constriction is usually symmetric although localized areas or bands of constriction have been reported.[9,10] The term effusive constrictive pericarditis, popularized by Hancock, is used to refer to cases in which constrictive pericarditis coexists with a tense pericardial effusion.[11]

History

Descriptions of diseases of the pericardium have a long history and date back as far as Egyptian and Greek civilizations. One of the early descriptions of CP was by Richard Lower in 1669 who described a patient with dyspnea and an intermittent pulse. Kussmaul, in 1873, coined the term pulsus paradoxus for the phenomenon of an intermittent loss of pulse during inspiration despite still hearing the heartbeat. In 1896, the term Pick's disease was given to patients who had ascites and hepatomegaly (pseudocirrhosis) in the setting of CP. See Table 4 for a summary of major historical events.[2,12]

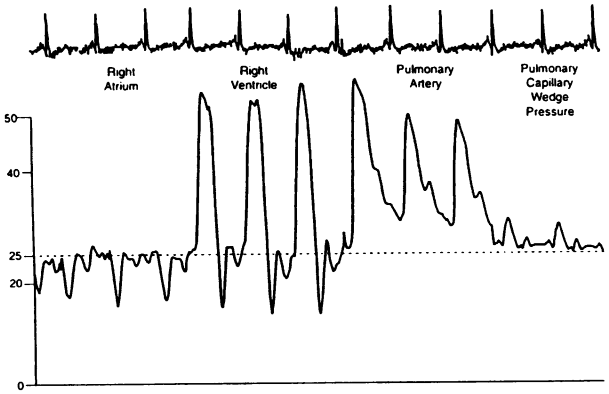

Figure 1.—Right atrial, right ventricular, pulmonary artery, and pulmonary capillary wedge hemodynamic tracings from a patient with CP. Note the diastolic equalization of pressures (less than 5 mm difference) and the pronounced x and y descent of the right atrial tracing.

Pathophysiology

The normal pericardium—a strong, thin sac surrounding the heart with fibrous tissue extending to the adventitia of the great vessels—is composed of two layers: a tough, fibrous layer (the parietal pericardium) and a smooth, serous layer (the visceral pericardium) which is intimately apposed to the heart's surface and epicardial fat. The normal visceral and parietal pericardium allow the heart to move around freely with uninhibited expansion of the cardiac chambers during diastole. This expansion provides for an unimpeded atrial contribution to diastolic filling, which has been reported to increase cardiac output by as much as 30%. With CP, however, the thickened, fibrotic pericardium has a relatively fixed diastolic volume which does not allow for

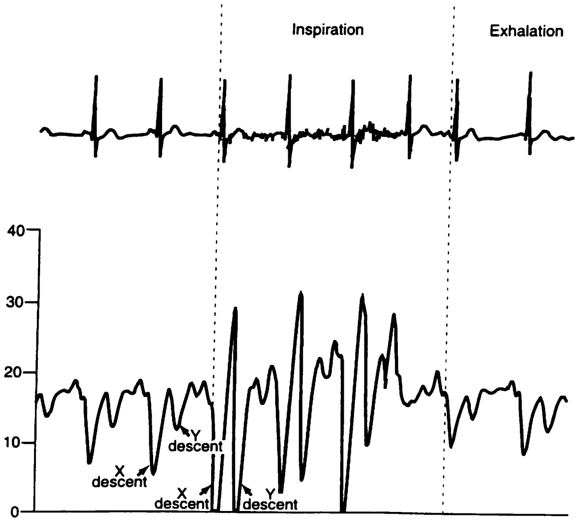

Figure 2.—Kussmaul's sign on hemodynamic tracing. Note the exaggerated x and y descent particularly during inspiration.

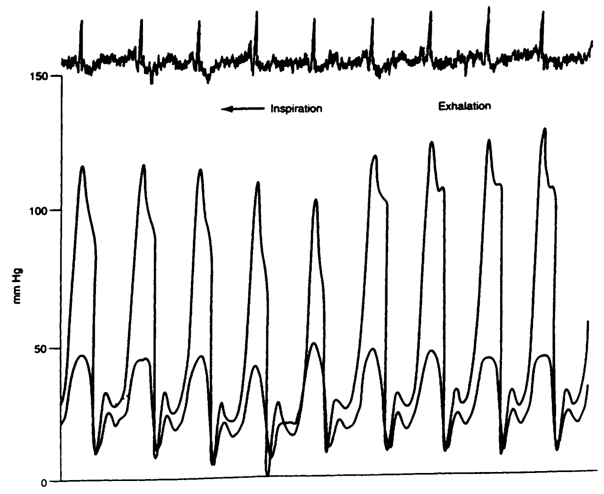

Figure 3.—Simultaneous right ventricular and left ventricular tracings. Note the close approximation of diastolic pressures.

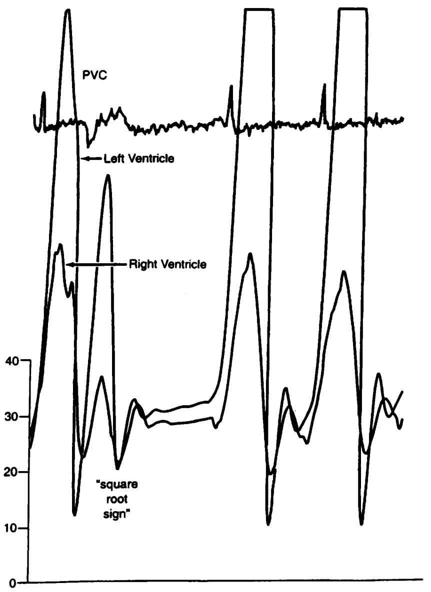

Figure 4.—Simultaneous right ventricular and left ventricular tracings. Note the near equalization of diastolic pressures, the rapid filling wave (√) which are most pronounced following a premature ventricular contraction.

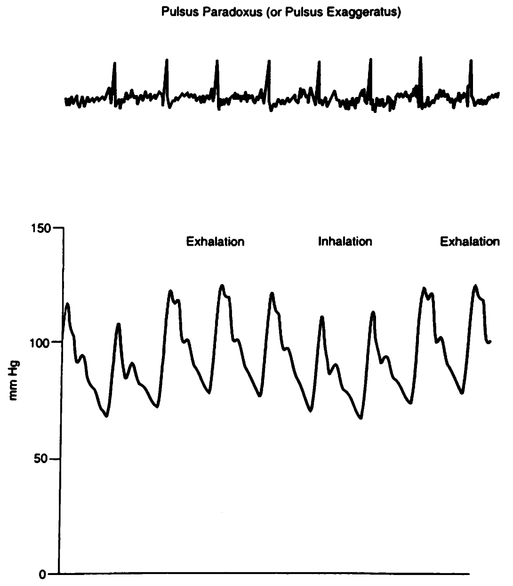

Figure 5.—Aortic pressure tracing demonstrating a greater than 15 mm respiratory change in systolic pressure consistent with pulsus paradoxus (or pulsus exaggeratus).

complete unimpeded filling. The cardiac chambers fill freely in early diastole because the volume in the cardiac chambers is low. But, as the cardiac chambers rapidly fill, and the volumes reach the threshold set by the thickened noncompliant pericardium, diastolic filling is abruptly halted. This sudden cessation may contribute to the exaggerated effect of ventricular interdependence (or coupling) with relatively more blood in the right atrium and right ventricle than in the left sided chambers during inspiration (Figure 6).

Pressure tracings of ventricular filling in patients with CP show that nearly all ventricular filling occurs in the first third of diastole with little filling after this because of the restraining effect of the thickened pericardium. This drop in activity accounts for the rapid y-descent and plateau seen in the JVP on physical exam, and in the right atrial, right ventricular and left ventricular pressure tracings during cardiac catheterization. The rapid y-descent represents the pressure in initial unimpeded chamber filling of early diastole, whereas the plateau represents the pressure at the fixed diastolic volume of mid- and late diastole as determined by the thickened pericardium. This dip and plateau on hemodynamic tracings is known as the "square root sign" (Figure 4).

The fixed diastolic volume determined by the thickened pericardium causes both right and left heart diastolic pressures to be elevated to the same degree (within 5 mmHg) in all four cardiac chambers. Thus, right atrial pressure, right ventricular diastolic pressure, and pulmonary capillary wedge pressure which indirectly represents left atrial and left ventricular diastolic pressures may all be nearly the same during right heart catheterization, a condition often referred to as diastolic equalization of pressures (Table 5). Furthermore, simultaneous right and left ventricular pressure tracings reveal almost a superimposition of the diastolic pressure tracings (Figures 3 and 4).

The mechanism of Kussmaul's sign is still debated, although one explanation is that the rigid pericardium in CP fails to transmit intrathoracic pressure variations to the heart's chambers.[13] Because the rigid pericardium in CP

TABLE 1.—*Laboratory Results*

Urinalysis: normal with no protein			
Alkaline Phosphatase: 153		SGOT: 26	SGPT: 18
Total Bilirubin: 1.9	Albumen: 3.9	Protime: 11.8 (INR = 1.0)	
BUN: 13	Creatinine: 1.1	WBC: 7.3	Hemoglobin: 15.5
Hematocrit: 45.9%	Platelets: 226,000	MCV: 86	

CXR: Normal cardiac sillhouette without pulmonary venous congestion
EKG: Normal sinus rhythm with borderline low voltage.
Echocardiogram: Biatrial enlargement, normal valvular function, and normal right ventricular and left ventricular systolic function with evidence of pericardial thickening. The Doppler mitral inflow pattern was suggestive but not diagnostic of constrictive physiology.

TABLE 2.—*Liver Function Tests: Passive Congestion vs. Cirrhosis*

	RV Failure	*Cirrhosis*
T. Bili.	↑	↑↑
Alk. Phos.	↑↑	↑
AST/ALT	usually nl	↑↑
Albumen	usually nl	↓
PT/INR	↑ or nl	↑↑

TABLE 3.— *Prevalence of Various Physical Examination Findings*

- Pulsus paradoxus usually absent (14-84%)
- Jugular veins distended (99-100%) - prominent y-descent
- Kussmaul's sign
- Pericardial knock/S3 (5-55%)
- Hepatomegaly (70-100%)
- Splenomegaly
- Ascites
- Peripheral edema (64-80%)

does not allow for right atrial and right ventricular expansion during inspiration, the increased venous return that occurs in inspiration results in a rise in central venous pressure. Another proposed explanation is that there is a normal inspiratory increase of intra-abdominal pressure transmitted to a tense overly filled venous system caused by CP.[14] Figure 6 is a schematic drawing demonstrating the mechanism of pulsus paradoxus (the drop in arterial pulse pressure during respiration [Figure 5]), a sign often seen in CP. Occasionally, cases of CP are seen without a pulsus paradoxus since the thickened pericardium can completely isolate the heart from the effects of respiration.[4]

Etiology

Tuberculosis is still the leading cause of CP in developing countries, but accounts for only a minority of cases in western cultures.[15] Most cases of CP in developed countries have no known antecedents although the possibility has been suggested that these cases are stem from an apparent previous episode of viral pericarditis.[3]

TABLE 4.—*Major Historical Events in Constrictive Pericarditis*

- 1669 Richard Lower describes a patient with dyspnea and intermittent pulse.
- 1842 Corrigan describes the pericardial knock (bruit de frappement).
- 1873 Kussmaul names the "paradadoxical arterial pulse," pulsus paradoxus.
- 1896 The eponym Pick's disease is given to patients with constrictive pericarditis who have ascites and hepatomegaly (pseudocirrhosis).
- 1929 The first successful pericardiectomy in the United States is performed by Ed Churchill.
- 1935 Paul Dudley White describes 7 of 15 patients with constrictive pericarditis who were successfully operated on at the Massachussetes General Hospital.
- 1946 Bloomfield demonstrates elevated right ventricular pressure with early dip and plateau in patients with constrictive pericrditis.
- 1982 Isner demonstrates the value of computed tomography in the diagnosis of constrictive pericarditis.

The frequency of radiation therapy as a cause can be influenced by the patients recruited to the series. For example, post-radiotherapy was cited as a cause of CP in 31% of patients in a Stanford series and was the etiology in only 5% in a Mayo series.[8,16] This discrepancy likely reflects the fact that Stanford is a large referral center for patients with Hodgkin's disease who often receive mediastinal radiation. The list of etiologies of CP are listed in Table 6.[2-4]

Diagnostic Evaluation

Routine studies such as ECG and chest radiographs usually are helpful only in supporting a search for pericar-

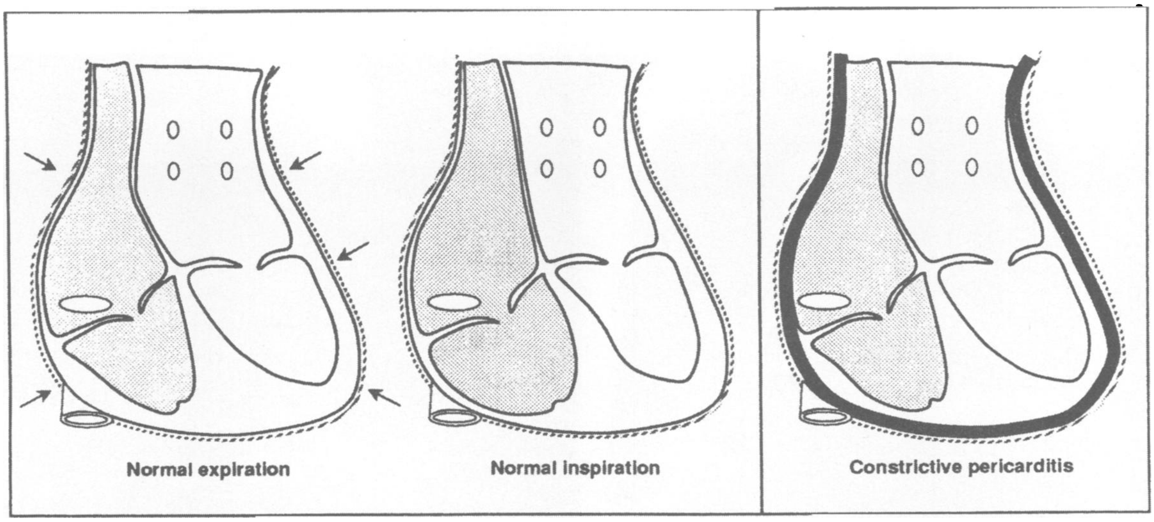

Figure 6.—Schematic drawing of the proposed physiology of pulsus paradoxus. Note the effects of inspiration on the right atrium and right ventricle in the normal heart causing bowing of the interventricular septum and a resultant decrease in left ventricular preload and therefore a decreased pulse. In constrictive pericarditis, however, a thickened and rigid pericardium can prevent transmission of intathoracic pressure variations to the right atrium and right ventricle resulting in little effect of respiration on the intracardiac pressures and hence little pulse pressure variation with respiration.

TABLE 5.—*Causes of Diastolic Equalization of Pressure**

Constrictive pericarditis
Pericardial tamponade
Restrictive cardiomyopathy
End stage dilated cardiomyopathy (All pressures high)
Dehydration (All pressures low)
Atrial septal defect
Hyperinflated lungs (COPD, pneumothorax)

*Less than 5 mm difference between right atrial and left atrial (pulmonary capillary wedge pressure), right ventricular and left ventricular diastolic pressure.

dial constriction and not for making a diagnosis. The findings on ECG, though rarely normal, are nonspecific and highly variable. Atrial arrhythmias frequently are seen with atrial fibrillation being a common late occurrence. Low voltage is seen less than 50% of the time, and left atrial enlargement is variable, ranging from 19-37% of cases.[17] The chest radiograph can reveal an enlarged cardiac silhouette, and pulmonary venous congestion and pleural effusions often are seen late in the disease. A calcified pericardium seen on a chest radiography is highly suggestive of CP when it is present in a patient with constrictive/restrictive physiology (Figure 7).[18] With the previous exceptions, the signs on ECG and chest radiographs are insensitive indicators of pericardial constriction. Cardiac catheterization often is required to document restrictive/constrictive physiology, and special imaging studies may be needed to prove the presence of a thickened pericardium.

Echo Doppler

Numerous non-specific echocardiographic findings in CP also may be found individually in other diseases.

TABLE 6.—*Major Categories of Etiologies of Constrictive Pericarditis*

1. Idiopathic
2. Infectious:
   - Tuberculosis
   - Viral esp. Coxackie B
   - Bacterial
   - Fungal
   - Parasitic
3. Post radiotherapy
4. Post cardiac surgery
5. Post traumatic
6. Neoplastic
7. Connective tissue diseases
   esp. rheumatoid arthritis and SLE
8. Toxic/Metabolic
   uremia, chylous pericardium, methysergide
9. Post myocardial infarction
10. Familial

Although echocardiography is insensitive in determining pericardial thickening, certain echocardiographic signs present in the appropriate clinical setting may argue strongly for the diagnosis of CP.[10] Often these echo findings are consistent with the physiology of CP: early rapid diastolic filling of the ventricles with little distension late in diastole because of a thick fibrous indistensible pericardium. Right and left ventricular systolic function is usually normal, however, because right and left ventricular cavity dimensions generally are preserved and systolic wall motion is normal. High atrial pressures may lead to right atrial and left atrial enlargement, inferior vena caval plethora, and prominent early diastolic ventricular filling causing exaggerated septal and posterior wall motion filling pattern.[19,20] The exag-

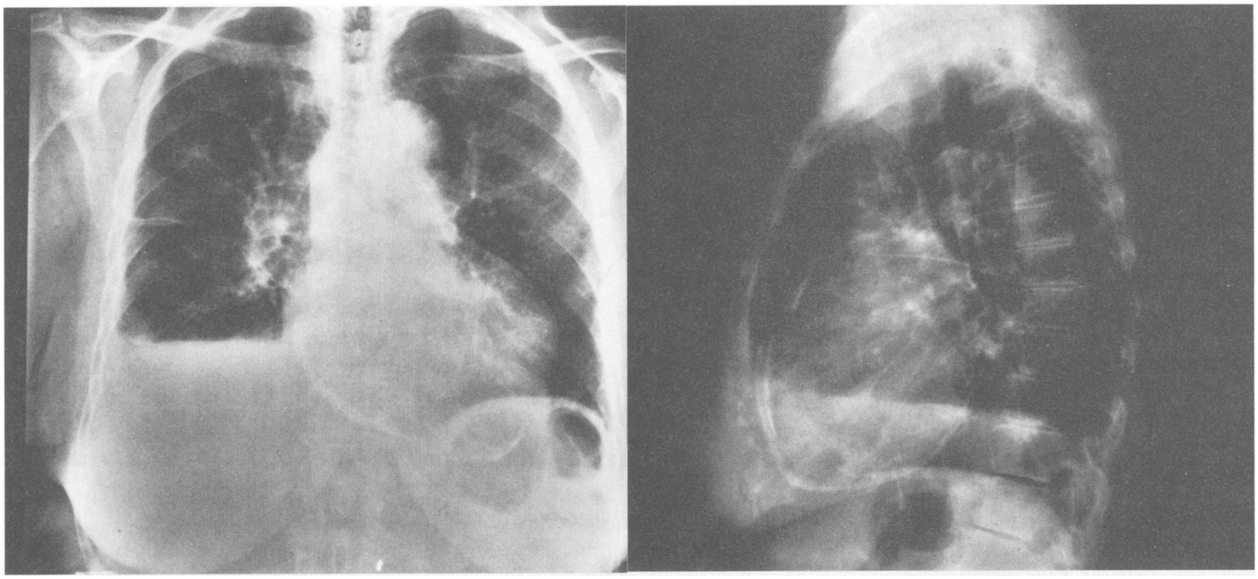

Figure 7.—Posterior-anterior and lateral chest radiograph demonstrating a thickened calcified pericardium in another patient with CP (Courtesy of Paul Stark).

TABLE 7.—*Echocardiographic and Doppler Signs of CP*

M-mode and Two D echo:

- Pericardial thickening
- Normal RV and LV chamber size
- Lefl atrial and right atrial enlargement
- Abnormal septal motion and posterior wall motion Paradoxical septal motion Diastolic septal bounce
- Premature opening of the pulmonic valve
- Dilated inferior vena cave without respiratory variation

Doppler Findings:

- Superior Vena Cava: Diastolic Flow Velocity > Systolic Flow Velocity
- Hepatic Vein: Exaggerated diastolic flow reversal afler onset of exhalation
- Pulmonary Venous Flow: Increase in diastolic flow velocity on exhalation Systolic/Diastolic flow velocity ratio < .65 Peak Diastolic flow velocity falls 40% on inspiration
- Mitral Inflow Pattern: During onset of exhalation 2 25% increase in early diastolic flow velocity

TABLE 8.—*Causes of Pulaus Paradoxus (Pulsus Exaggeratus)*

- Pericardial Tamponade
- Exaggerated Respiratory Motion
- Asthma
- Decompensated Obstructive Lung Disease
- Effusive Constrictive Pericarditis
- Constrictive Pericarditis
- Restrictive Cardiomyopathy

gerated response to respiration may lead also to abnormal atrial and ventricular septal motion seen on routine transthoracic echo but often is best noted on transesophageal images.

Adding a new dimension to the evaluation of diastolic function, Doppler echocardiography has become extremely useful not only in diagnosing CP but differentiating it from restrictive cardiomyopathy. Again, many of the findings may be nonspecific and are found in patients either with exaggerated breathing patterns of pulmonary disease or with right ventricular infarction. Some findings are listed in Table 7, and more technical explanations of the mechanisms for these findings can found in References.[19, 21-24, 29]

Hemodynamics

The hallmark of the hemodynamic evaluation of CP is diastolic equalization of pressure (less than a 5 mm difference between right and left atrial pressures and right and left ventricular diastolic pressures) at an elevated level. The encasement of the heart by a thick non-compliant pericardium as noted earlier leads both to this equalization of pressure and to a lack of transmitted intrathoracic pressure changes upon the heart. The vascular structures in the thoracic cavity (the superior and inferior vena cava) continue to be affected by the respiratory changes in the thoracic cavity. Consequently, inspiration allows more filling of the right atrium and right ventricle causing increased right atrial and right ventricular septal impingement on the left atrial and left ventricular cavities which have received less venous filling because of pulmonary venous pooling during inspiration (Figure 6).[25] During exhalation the reverse occurs. Exaggerated inspiratory right atrial pressures and filling patterns referred to as Kussmaul's sign may be noticeable on the pressure tracings (Figure 2) . Less frequently, one may note also an exaggerated difference between aortic systolic pressure during inspiration and exhalation, historically called "pulsus paradoxus" but more appropriately called "pulsus exaggeratus" because it is an exaggeration of a normal physiologic phenomenon of a decrease in pulse pressure with respiration (Table 8). Figure 1 shows the hemodynamic tracings of this patient with CP, demonstrating diastolic equalization of pressure.

Other tracings from this same patient demonstrate the similarity of the diastolic pressures of the right and left ventricle. Often this pressure similarity may be made clear after a premature ventricular contraction or during inspiration (Figures 3 and 4). If the heart rate is slow enough, a "√" configuration may be noticeable because of the exaggerated rapid filling wave and pressure plateau secondary to lack of further ventricular filling. In an attempt to differentiate CP from restrictive cardiomyopathy, physicians have used other criteria, such as the ratio of right ventricular diastolic pressure to systolic pressure of less than 1 to 3 and right sided systolic pressures of less than 55 mmHg found more commonly in CP (Table 9).[26,27] None of these hemodynamic findings alone are diagnostic of CP but only consistent with it. All of the numerous other causes of diastolic equalization of pressure, as noted in Table 5, should be excluded before diagnosing CP. Restrictive cardiomyopathy with amyloidosis is the most likely to simulate CP.[27,28] In cases where all of the diastolic pressures remain low and there is a suspicion of CP, a one-liter intravenous fluid bolus may be given to look for continued diastolic

TABLE 9.—*Hemodynamic Catheterization Findings of Constrictive Pericarditis*

1. A left ventricular diastolic and right ventricular diastolic pressure difference of less than 5 mmHg at rest. This becomes more obvious post PVC and one beat after onset of inspiration.
2. Diastolic dip and plateau (square root sign) of ventricular tracing. Prominent rapid filling wave.
3. Lack of respiratory variation or an increase in right atrial pressure on inspiration (Kussmaul's sign).
4. Right ventricular end-diastolic pressure to right ventricular systolic pressure ratio greater then 1/3.
5. Right ventricular systolic or pulmonary artery pressure greater than 55 mmHg.

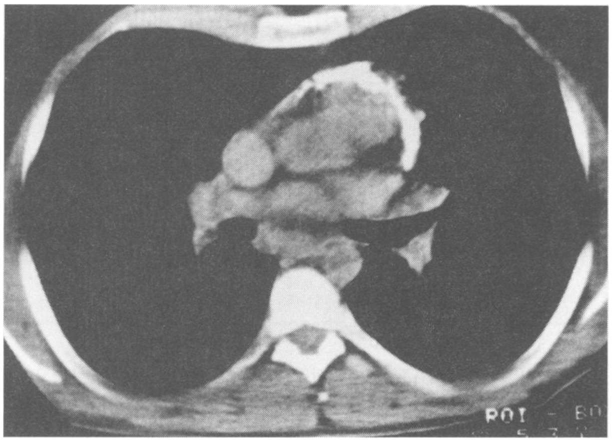

Figure 8.—Computed tomography of the chest in another patient with CP demonstrating a thickened pericardium (>4 mm.) (Courtesy of Paul Stark).

equalization of pressure (so-called occult constrictive pericarditis).[30] In normal dehydrated patients, the right and left ventricular diastolic pressures will begin to separate by more than 5 mmHg after fluid challenge. In cases of CP, the right and left ventricular diastolic pressures may rise but will not separate.

Imaging Modalities

The enhanced resolution of radiologic imaging has improved the sensitivity in detecting evidence for pericardial constriction. Computed tomography (CT) and magnetic resonance imaging (MRI) showing a thickened pericardium (Æ4 mm in most studies) in a patient with a hemodynamic profile consistent with constrictive/restrictive physiology is considered diagnostic of CP until proven otherwise (Figure 8).[12,31-35] The sensitivity of CT and MRI, however, range from 75-100%, and many cases of CP have been reported in patients whose pericardium is of normal thickness.[2] Therefore, pericardiectomy still may be the only way to confirm the diagnosis in patients whose hemodynamic profile suggests constriction but imaging is unable to document pericardial thickening, and a diagnosis cannot be confirmed by endomyocardial biopsy.[37]

Treatment

Although relatively asymptomatic patients can be managed initially with diuretics, particularly if the diagnosis is not as clear cut as expected, pericardiectomy with complete decortication of the ventricular pericardium is the treatment of choice for CP. The left anterolateral thoracotomy approach is preferred by some surgeons, although most prefer to do a median sternotomy with or without cardiopulmonary bypass.[35] This approach permits greater mobilization of the heart and allows for complete removal of the ventricular pericardium, including the entire diaphragmatic pericardium.[5,16,38] In some cases of CP, constrictive physiology is still found postoperatively despite complete removal of the parietal pericardium. Reoperation with removal of a fibrous epicardium has proven successful in some of these cases with a "constrictive epicardium," but is hazardous to the patient because of the proximity of the epicardial vessels.[5,8] Failure to improve postoperatively may result from inadequate decortication of all fibrous parietal and visceral pericardium. Other factors, such as myocardial atrophy and progressive ventricular dilation, may result in postoperative heart failure.[16] In hemodynamics, days or weeks of cardiopulmonary support often are required before gradual improvement is accomplished.

The operative mortality is highly dependent on the preoperative NYHA functional class status. In a series of 313 patients at the Mayo Clinic (from 1936-1990), the operative mortality was 1% for patients in class I or II, 10% for patients in class III, and 46% for patients in class IV status, with an overall mortality of 14%.[5] Similar mortality rates for patients in the different NYHA classes have been reported in other series with overall mortality rates ranging from 5-15%.[6,16] Other preoperative risk factors include degree of right ventricular end diastolic pressure, diuretic use, renal insufficiency, and the presence of radiation pericarditis. Risk factors found intraoperatively that worsen prognosis include the presence of unresectable calcifications or incomplete pericardial decortication.[5,6,8,16]

In general, patients with CP have a good outcome when their disease process is discovered early in its course.[6,38] Physicians must be able to recognize the subtle differences in clinical presentation of patients with CP, restrictive cardiomyopathy, pericardial tamponade, right ventricular failure, and chronic liver disease. Careful physical examination with close attention to the neck veins, a cardiac exam, and determination of liver size and texture with the appropriately directed diagnostic studies are important for recognizing this rare disease. Since surgery often results in a dramatic improvement in symptoms with the best prognosis in patients with early disease, cardiology referral for cases of suspected CP is most important. Given the highly curable nature of this disease, in cases where constrictive/restrictive physiology is present and imaging studies do not demonstrate a thickened pericardium, cardiothoracic surgery referral for diagnostic and therapeutic pericardiectomy still may be warranted.

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